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A forgotten disease: Superior mesenteric artery syndrome

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Abstract

Aim: Superior mesenteric artery syndrome (SMAS) is a rare cause of upper gastrointestinal obstruction. As it is encountered infrequently, there have been only a few studies on it, mostly in the form of case series or just a case presentation in the literature. In this study, we aim to investigate common clinical features and radiological measurements between patients admitted with symptoms of gastric outlet obstruction due to SMAS and those without SMAS. Material and Method: This study was conducted prospectively on individuals admitted to our clinic presenting with symptoms of gastric outlet obstruction in the previous six months and who had undergone computerized tomography. (Here, SMAS is defined by a narrow aortomesenteric angle and a short aortomesenteric distance.) Demographic features and radiological measurements were compared between patients with and without SMAS. The results of a three-month follow-up of each subject with SMAS were recorded. Results: This study includes a total of 92 subjects, 14 (15%) of whom had a diagnosis of SMAS (Group 1) and 78 (85%) of whom did not (Group 2). Group 1 had a predominance of female and significantly younger patients (mean age: 31.1 ± 10.2), compared to Group 2 (mean age: 44 ± 11.7) ($p < 0.001$). Abdominal subcutaneous fat tissue thickness was thinner in group 1 than in group 2 (18.1 ± 8.1 and 23.7 ± 12.2, respectively; $p = 0.039$). Eleven of the 14 SMAS patients (78.5%) recovered with medical treatment, but in the other three cases, surgical intervention was considered. Discussion: SMAS is an important and preventable cause of small bowel obstruction. Although it is a rare condition in the general population, accurate diagnosis of the disease leads to improvement of symptoms; simple medical management can preclude the need for surgery.

Keywords

Intestinal Obstruction; Wilkie's Syndrome; Superior Mesenteric Artery Syndrome; Endoscopy; Vomiting; Anorexia; Computerized Tomography

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Introduction

Superior mesenteric artery syndrome (SMAS), also known as Wilkie's syndrome, is a rare condition characterized by vascular compression of the duodenum. It results from occasional loss of the mesenteric fat pad, in the setting of rapid weight reduction due to a several conditions (e.g. tuberculosis, anorexia nervosa, abdominal trauma, burns, scoliosis surgery, or a high insertion of the ligament of Treitz [1–4]). In such cases, the aortomesenteric angle (AMA) is narrowed and it presents a partial small bowel obstruction due to compression of the third part of the duodenum, between the superior mesenteric artery and the aorta [5].

SMAS was first described by von Rokitansky, in 1842 [6,7]. However, the disease had been better defined by Wilkie, in clinical and pathophysiological terms; following a series of 64 patients, he identified this condition as chronic duodenal ileus. He also suggested treatment approaches [8,9]. Reviewing the literature, there have been only a few studies mostly case series, or a case presentation. In addition, no randomized clinical trial has been performed regarding the management of patients with SMAS. Therefore, there is no consensus on SMAS diagnostic work-up or management approaches.

In the current study, we aim to investigate common clinical features and radiological measurements between patients admitted with symptoms of gastric outlet obstruction due to SMAS, and those without SMAS and pointed out of the clinical importance of this disease is rare but might be mortal.

Material and Method

This cross-sectional prospective study was approved by the institutional ethical board, and accords with the Helsinki Declaration. This study was conducted on individuals admitted to our clinic with one or more of the clinical signs of gastric outlet obstruction namely, weight loss, postprandial dyspepsia, and recurrent vomiting with or without chronic or severe acute on chronic abdominal pain in the previous six months, and who had undergone computerized tomography (CT) and upper gastrointestinal endoscopy through appropriate indications between October 2013 and October 2015 at a single centre in Turkey.

The patients were provided written information about the study and asked to complete a standard questionnaire asking about their personal medical history, present medications, family history, and lifestyle habits. Data such as their clinical presentation, how their diagnosis was substantiated, the offered treatment, and their response to the treatment were collected and recorded. The exclusion criteria were: age <18 years, pregnancy, systemic infection, any thyroid or liver disease, impairment of kidney function, history or diagnosis of colorectal disease, gastrointestinal malignancy, or prior gastrointestinal surgery.

The aortomesenteric angle (AMA), aortomesenteric distance (AMD), and subcutaneous fat tissue thickness (SCFTT) of all cases were measured from CT images. Subjects with a narrow AMA (<35°) (Figure 1) and an AMD <8 mm (Figure 2) were diagnosed as having SMAS (10). Cases with SMAS comprised Group 1; those without SMAS comprised Group 2. The two groups were compared in terms of demographic characteristics and all radiological measurements.

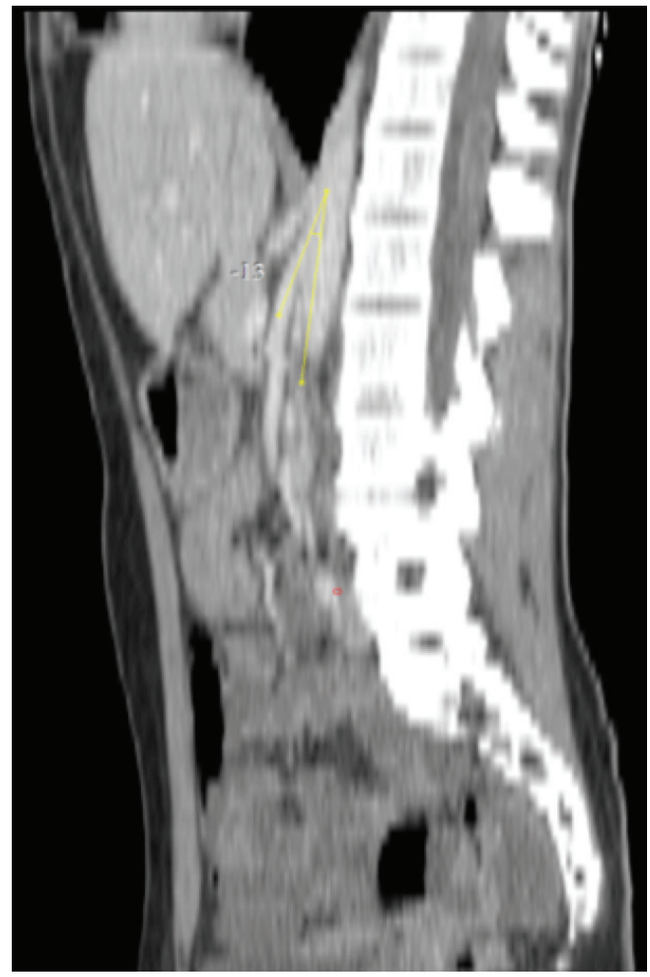

Figure 1. Computerized tomography scan of the abdomen showing narrowed Aorta mesenteric angle

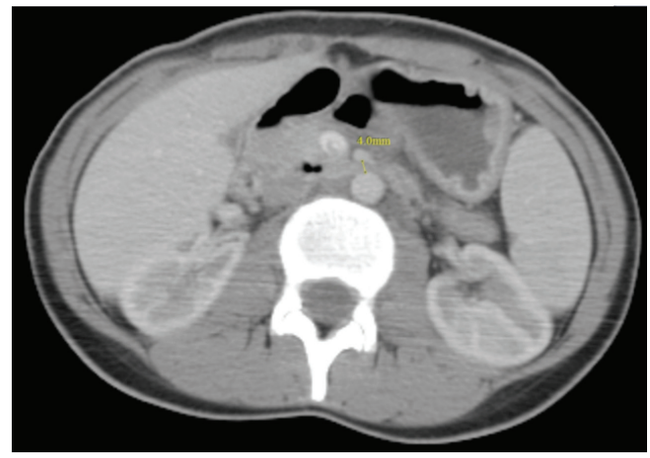

Figure 2. Computerized tomography scan of the abdomen showing narrowed Aorta mesenteric distance

A number of medical managements had been recommended to all SMAS cases, including a knee-chest or side-lying position after eating; the placement of a nasogastric tube, through which approximately 2,000 mL of stomach contents could be drained; and the administration of a proton pump inhibitor. In cases in which a patient's symptoms had not been resolved by the initial treatment, a nasojejunal tube was inserted. Medical management was planned for a two-week period, but the duration of medical treatment was extended to one month for patients whose symptoms had not completely improved.

Cases unresponsive to the medical approaches underwent surgery. We recorded a follow-up for each subject in Group 1 three months after the treatment.

Statistical Analysis

Data analysis was performed using the statistical software package programme IBM SPSS software for Windows (version 19.1; SPSS Inc., Chicago, IL, USA). Intergroup comparisons of categorical variables were done using a chi-square test, and continuous variables were compared using a student *t*-test. Categorical variables were presented as percentages or counts, and continuous variables were presented as a mean ± standard deviation. A value of $p < 0.05$ was considered statistically significant.

Results

This study includes a total of 92 subjects who were admitted with one or more of the clinical signs of gastric outlet obstruction, 14 (15%) of whom had been diagnosed with SMAS (Group 1) and 78 (85%) of whom did not have SMAS (Group 2) as we have previously described.

The demographic features and radiological measurements of all groups are presented in Table 1. There was a predominance of female patients in Group 1 (78%; n = 11); in Group 2, they comprised 39% (n = 30) ($p < 0.001$). Group 1 had significantly younger patients than did Group 2 (mean age, 31.1 ± 10.2 and 44 ± 11.7, respectively; $p < 0.001$). Abdominal SCFTT was significantly thinner in Group 1 than in Group 2 (18.1 ± 8.1 and 23.7 ± 12.2, respectively; p = 0.039) (Table 1).

The demographic features and radiological measurements of each patient in Group 1 are presented in Table 2.

As for the frequencies of the symptoms of the groups, weight loss symptoms were more common in Group 1 than Group 2.

Whereas, endoscopic findings of 11 patients were normal, endoscopic findings of just 3 patients were abnormal which was dilated bulbus.

Eleven patients symptoms were improved by medical treatment, only three patients need the surgery but one of them refused the surgery.

Discussion

We found that 14 (15%) of the patients who presented to our hospital with clinical signs of gastric outlet obstruction were diagnosed as SMAS; a majority of those SMAS patients responded to medical treatment. These findings indicate that SMAS is an important cause of small bowel obstruction, and a sizeable proportion of cases could be prevented through medical management.

Von Rokitansky reports that SMAS has an incidence of 0.013–0.3% in the general population, with a mortality rate of 33%. Considering that our hospital is a 450-bed hospital that serves a population of 300,000 individuals, this one-year record of SMAS cases translates into an incidence rate of 0.05%. This result aligns with the findings in the literature [6]. Although it is a rare condition in the general population, it is important as a cause of small bowel obstruction that can be prevented through medical management. Precisely for this reason, this diagnosis should be kept in mind in patients presenting with intestinal obstructive symptoms.

Patients with SMAS present several nonspecific symptoms, either acutely, or chronically with acute exacerbations; these include: vomiting, epigastric pain, postprandial discomfort, and weight loss [11,12]. Symptoms usually occur after, or are aggravated by, eating; they are relieved by postural changes, such as turning onto the left side, in either a prone or knee-chest position [13]. In this study, revealed that vomiting was the most common symptoms, followed by weight loss in SMAS patients.

Although the clinical manifestations of SMAS are shared with many of the common causes of gastric outlet obstruction, this condition has unique radiological features. In humans, the aorta SMA angle ranges from 38° to 70°, depending on body posture [1]. The main radiological feature of SMAS is a narrowing of the aorta SMA angle, to within a range of 1–35°; therefore, the AMD decreases to <10 mm, from a typical distance of 10–28 mm [14]. In addition, CT may show dilatation of the stomach and duodenum down to the third part, with a sudden cut-off distally, conforming to the anatomical position of the superior mesenteric artery. Moreover, endoscopically gastric or duodenal dilatation might be seen, but special attention is needed to diagnose dilatation. In our case series, we diagnosed dilatation of the duodenum only in three patients.

Table 1. Demographic and radiologic features of both two groups.

	Group 1	Group 2	p
Age	31.1 ± 10.2	44 ± 11.7	<0.0001
Sex (female)	78% (n:11)	39% (n:30)	<0.0001
Most common presentation	Vomiting, Weight Loss	Vomiting, Postprandial Dispepsia	
AMA	21.4±6.8	50.1±18.4	<0.0001
AMD	5.2±1.2	15.1±7.1	<0.0001
SCFTT	18.1 ± 8,1	23.7 ± 12.2	0.039

Abbrevations: AMA: Aortomesenteric Angle, AMD: Aortomesenteric Distance, SCFTT: Subcutaneous Fat Tissue Thickness

Table 2. Demographic, clinical and radiological features of each patients with superior mesenteric artery syndrome.

Patient	Sex	Age	Symptom	AMA	AMD	SCFTT	FUE	Treatment	Result
1st	W	42	AP, V	18	5	21	ND	Medical	Improved
2nd	W	37	AP, PPD	21	4,5	23	ND	Medical	Improved
3th	M	49	WL, V	21	5	18	DB	Medical	Improved
4th	W	37	WL, PPD	16	5	20	ND	Surgery	Improved
5th	W	22	WL, V	22	3,5	32	ND	Medical	Improved
6th	W	18	WL, V	22	4	7	ND	Medical	Improved
7th	W	19	AP, V	17	5	12	ND	Medical	Improved
8th	W	27	WL, V	22	4,5	9	ND	Medical	Improved
9th	W	33	AP, WL, V	23	5,5	12	ND	Medical	Improved
10th	W	32	AP, V	20	5	18	DB	Surgery	Improved
11th	M	20	AP, V	14,5	6	12	DB	Medical	Improved
12th	M	41	AP, WL	18	7,5	7	ND	Medical	Improved
13th	W	23	V, PPD	23	5,5	12	ND	Medical	Improved
14th	W	22	AP, V	24	4	28	ND	Medical	Not accept to surgery

Abbrevations: AMA: Aortomesenteric Angle, AMD: Aortomesenteric Distance, AP:Abdominal Pain, DB: Dilated Bulbus, FUE: Findings with Upper Endoscopy, SCFTT: Subcutaneous Fat Tissue Thickness, ND: Normal Duodenum, PPD:Postprandial Distension, V:Vomiting, WL:Weight Loss

Symptoms related to small bowel obstruction and characteristic radiological features facilitate SMAS diagnoses. However, its diagnosis is typically challenging and often delayed, given its insidious presentation. The diagnosis requires a high index of clinical suspicion in an accurate clinical context and underdetailed radiological evaluation. For instance, before being admitted to our clinic, six patients within our SMAS group had been referred to the psychiatry clinic because of their recurrent vomiting, and another patient had been administered colchicine for Familial Mediterranean Fever. A delay in diagnosis can potentially lead to many complications, such as electrolyte imbalance, catabolic wasting, peritonitis, and gastric perforation; ultimately, even if these other complications have not occurred, a patient may experience a low quality of life. In this study, 11 of the 14 patients who were early diagnosed were treated with only medical treatment and their clinical conditions were improved.

Weight loss is considered a SMAS risk factor, as it depletes the retroperitoneal fat and lymphatic tissues, a condition that leads to vascular compression of the third part of duodenum [15]. In line with this theory, our study found that the SCFTT of the SMAS group was significantly thinner that of the non-SMAS group.

First-line SMAS treatments include conservative management. Decompression of the stomach and duodenum is advised, with the aid of a nasogastric tube. Feeding can be provided through a nasojejunal tube or parenteral nutrition, with or without postpyloric feeding, if possible [16]; when tolerated, oral feeding can be resumed. Nutrition helps build up the fat cushion between the SMA and aorta and, hence, could reverse the situation, as seen in 11 of our patients. In addition, as this condition lies in the root of the mesentery, positional changes such as assuming a knee-chest or side-lying position after eating tend to widen the aorta–SMA distance, and can partially relieve the degree of duodenal obstruction and facilitate the movement of gastric contents through the area of constriction [17]. No time limit has yet been defined for the medical treatment; in our study, the symptoms of eight patients improved within two weeks; six of them had been treated medically for one month. Finally, three of our patients did not respond to the medical treatment, and for them, surgical treatment was recommended. While one of them did not agree to the surgery, the other two did, and their symptoms improved thereafter.

With medical treatment, most patients eventually recover; however, in some cases, symptoms can worsen, and severe gastric dilatation that requires hospitalization may occur. In such cases, the cause often remains unidentified. In these cases, surgical intervention may be considered [18]. The aim of such surgery is to bypass the site of obstruction; therefore, in cases of SMAS, gastrojejunostomy and especially duodenojejunostomy are the procedures of choice. Duodenojejunostomy relieves the obstruction, and bears a success rate of up to 90%; it can be performed with open laparotomy, laparoscopy, or robotic duodenojejunostomy [18–20].

The main limitation of this study was that we did not measure AMA, AMD, or SCFTT by CT after treatment. We needed to avoid the use of radiation for this purpose, as this type of treatment was neither ethical nor cost-effective.

In conclusion, given the low incidence of SMAS, it can be easily overlooked. Although it is a rare condition in the general population, accurate diagnosis of the disease and subsequent treatment with simple medical management leads to improvement of small bowel obstruction; under such circumstances unnecessary surgery can be completely precluded. Conservative approaches to SMAS as first-line therapies are usually effective, and weight gain clearly correlates with relief of symptoms. Even in cases in which patients do not improve with conservative management, surgical treatment is rarely indicated. Therefore, SMAS is a curable disease when it is diagnosed in a timely fashion.

Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.

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Conflict of interest

None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

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